To the Editor:  
We read with great interest the report by Yoshigi et al, describing a patient with tetralogy of Fallot and co-ex-isting aortic coarctation. Because of the rarity of this asso-ciation, we describe a seventh patient, adding a few comments about possible pathogenesis.

A one-year-old girl with congenital hypothyroidism became increasingly cyanotic. Echocardiography and angiography revealed tetralogy of Fallot, a left aortic arch with mild tubular hypoplasia beyond the right common carotid artery, and small left superior caval vein. The left anterior descending coronary artery coursed anomalously over the infundibulum of the right ven-tricle. The gradient across the isthmus was 30 mm Hg.

Following resection of the coarctation and end-to-end repair, a modified Blalock-Taussig shunt was con-structed. The postoperative course was unremarkable. At the age of three, she underwent definitive correction of the malformation. The child is presently in good condition.

Cardiovascular malformations can be classified in many ways. One system is based upon postulated disordered embryonic mechanisms. In this mechanistic approach, tetralogy is attributed to abnormal migration of ectomesenchymal tissue, and loosely labelled the “conotruncal and branchial arch vessel” group. Coarctation is postulated to be due to altered intracardiac blood flow, and placed in a different grouping. It is possible, however, that certain cardiovascular malfor-mations, such as coarctation, may be mechanistically heterogeneous. If the theory of altered flow is correct, the most coarctations associated with the typical juxtaductal shelf would be considered a disruption of normal cardiac development. The anatomy in our patient, and the other patients reported in association with tetralogy, in contrast, has been described as involving a longer hypoplastic segment of the arch, and arrangement known as tubular hypoplasia. We specu-late that these cases may be viewed as a primary develop-ment error, and thus classified in a group other than that involving disordered flow. In other words, coarctation may look different when it results from different embryonic mechanisms.

Luca Rosti, Angela E. Lin, Alessandro Frigiola, Anna M. Calli

Address for correspondence:
Dr Luca Rosti, Pediatric Cardiac Surgery, Centro E Malan,
Ospedale San Donato, via Morandi 30, 20097 San Donato Milanese, MI, Italy
Tel. 39 2 5277 4511; Fax. 39 2 527 4717

References